

## ON OCULAR AFFECTIONS IN SYPHILIS OF THE BRAIN, WITH REPORT OF FIVE CASES.<sup>1</sup>

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IN two previous papers which I had the honor to present to this society (*Transactions Wis. State Med. Soc.*, 1891 and 1892, and *The Medical and Surgical Reporter*, July 29, 1893, and Nov. 25, 1893) I endeavored to describe the intimate connection between the eye and the brain. The optic path, the third, fourth, and sixth nerves run the greatest part of their length within the cranial cavity, so that they become largely exposed to encephalic diseases. Not only the variety and frequency of functional disturbances, owing to this arrangement, but also the possibility of inspecting, with the ophthalmoscope, the entrance of the optic nerve in the eye and the retina, give a special value to the eye symptoms in affections of the brain. Therefore it will not be surprising that in syphilis, which may invade the brain in all its structures in various fashions, only about 15 per cent. of the cases are without ocular symptoms (Uhthoff).

The literature of this field is abundant, but only in the last years, with the perfection of methods and the great progress in examining the brain and nerves anatomically and especially histologically, have the observations become more accurate. One of the latest works is Uhthoff's admirable paper, based on 100 cases of brain-syphilis with ocular symptoms, observed by himself, of which 17 came to a post-mortem examination, and on 150 autopsies, compiled from literature (*von Graefe's Archives*, vol. xxxix., 1 and

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2, and vol. xl., 1). In the following I purpose to speak of ocular affections in syphilis of the brain, with the report of five cases of my own observation. The examination in all these has been very careful, but for the sake of brevity I shall omit the negative results, and state the positive ones only.

CASE 1.—*Hemiplegia, hemianopia, etc., from basal gummous meningitis.*—A man of sixty-two years was paralyzed in June, 1891. Syphilitic infection seven years previously. September, 1891, I saw him for the first time in consultation on account of impairment of sight, and ascertained right hemiplegia (face, arm, leg) and right homonymous hemianopia. V.  $\frac{1}{2}$  $\frac{5}{6}$ . Ophth. normal. Tongue deviates to the right when protruded, to the left when drawn back. Defective articulation, speech often unintelligible, failure of memory, amnesic aphasia. The right lateral hemianopia sometimes was complete, sometimes there was only hemiachromatopia, *i. e.*, all appeared gray in the right half-field, or hemiamblyopia, *i. e.*, objects could be seen, but dimly only, and seemed to be smaller. For instance, when looking at me with either eye, the left side of my face, my left eye, and hand appeared to him smaller, as if they belonged to a smaller person. Antisyphilitic treatment without result. The condition grew gradually worse. Dec. 29th the patient was lying on the lounge perfectly apathetic; did not speak; incontinence of urine and fæces. The ophthalmoscope revealed in both eyes very marked papillitis with slight hemorrhages into the substance of the optic discs. Tortuous engorged veins. Outlines of discs veiled, which were rather swollen and prominent. Pupils medium-sized, react very sluggishly to light. Jan. 1, 1892: Pulse, 120. Excessive perspiration. Very restless. Right limbs painful when he is dressed. Jan. 6th, death, 9 P.M.

Jan. 7th, 1 P.M., I made the *autopsy*: The base of the left hemisphere is the seat of a diffuse growth, starting from the meninges, yellowish-gray, of lardaceous consistency, involving the frontal and temporal lobes, optic tracts, pons, middle and posterior cerebral arteries, and the third nerve. This growth is the result of thickening of the pia-arachnoid, due to syphilitic meningitis. It extends into the fissures and recesses and has grown into the walls of the vessels, conglomerating all these different portions of the brain into one mass. The trunk of the middle cerebral artery is dilated, and its coats are invaded by the gummous growth



which sheathes the artery all around. Its branches lead to a large focus of softening in the corpus striatum from arterial occlusion. Microscopically the growth at the base consists partly of fibrous tissue, and its portion at the frontal lobe is in a state of fatty degeneration, showing granule corpuscles, fatty granules, and detritus.

The features of interest in this case are the homonymous hemianopia and the papillitis. The former was caused by the affection of the left optic tract, which, with the frontal and temporal lobes, was the seat of the syphilitic new-formation, *i. e.*, it was due to a *basal* lesion. Indeed this basal gummous meningitis is the most common condition in syphilis of the brain. It may be diffuse or circumscribed, may be complicated with a tumor-like formation of gummous tissue, or a gumma proper may be the only affection. Or its seat may be the convexity of the brain.

Next in frequency is the syphilitic endarteritis (Heubner), and this may accompany the former. The softening of the brain at the corpus striatum was brought about by compression of the middle cerebral artery by the gummous neoplasm at the base, leading to thrombosis of its terminal twigs, and subsequent necrobiosis of the brain area supplied by it.

The fibres of the visual path are implicated very frequently (in fourteen out of Uhthoff's seventeen autopsies), but the optic tract, as in our case, rather exceptionally, possibly on account of its situation not being so much exposed. The seat of predilection and the starting-point of the gummous affections of the cranial base is undoubtedly the chiasm, which may be entirely imbedded in a gummous mass. This may extend downwards to the optic nerves, leading to a gummous degeneration with considerable swelling, or to optic neuritis or papillitis and perineuritis, with consequent neuritic atrophy or to descending atrophy. Quite a number of post-mortem examinations showed this condition, or gummous masses sheathing the optic nerves, not entering into their substance, without functional or ophthalmoscopic changes during life. Thus we cannot exclude its existence even in cases without corresponding clinical symptoms, if the diagnosis of cerebral syphilis is established by other

signs. Mostly it is not a primary gummous process in the optic nerve, but an interstitial affection invading the nerve from its periphery along the septa and replacing its fibres. The *intracerebral* optic fibres are much more rarely damaged than the basal (Uhthoff, xxxix., 1, p. 121).

The visual path may, however, be affected by way of the arteries supplying them. Endarteritis obliterans of the ophthalmic artery is of very rare occurrence (Uhthoff), and in case of obstruction its many anastomoses may prevent blindness by re-establishing the blood supply. Whenever it was observed, the functional disturbances could more easily be explained by direct gummous degeneration of the optic nerves. But endarteritis of the cerebral arteries proper is very frequent. Our second case shows the course of this affection.

CASE 2.—*Right hemiplegia, hemianæsthesia, etc. from syphilitic obliterating endarteritis of the left middle cerebral.*—A man, æt. forty-one, acquired syphilis 1887. In January, 1890, he suffered from severe headache day and night for weeks. He was treated at the hospital for one week, and rested at home for two weeks. The headache continued, however, more or less. July 15, 1890, his physician sent him to me for a laryngoscopic examination, as he complained of severe pain at the left side of the throat in the region of the fifth vertebra. Larynx healthy, but a very large and deep syphilitic ulcer at naso-pharynx. He looked careworn and emaciated; had lost eight pounds within a few weeks. I ordered large doses of iodide of potassium and treated the ulcer successfully.

Aug. 19th he was led into my office, having met with a paralytic stroke on the way, resulting in right hemiplegia (face, arm, leg). He said that the night before he suddenly felt numb in right side of body, with formication in his right limbs. But he got over it and slept well. This morning he went to his store perfectly well, and when he came to me he met with this accident. Sensibility lost on right side of face and body. He can raise his right arm only slowly and with great effort, but it soon falls down. His right leg is dragged behind and he cannot stand on it. Speech hesitating and in some words not clear. After sitting down and resting awhile he can walk and move his arm better, but he is weak. Sensibility returns. At once sent to hospital



and most energetic antisyphilitic treatment instituted. Aug. 27, 1890, walks pretty well and is improving rapidly. After three weeks at hospital he recovered so far that he was able to work as a clerk. I last saw him on April 7, 1894; no relapse in these four years. His weight increased from 121 pounds to 153. Except occasional paræsthesia in right leg and arm he feels perfectly well.

*Diagnosis:* Syphilitic endarteritis obliterans of left middle cerebral artery, narrowing its lumen and impeding and arresting the circulation. Fortunately only an ischæmia took place without thrombosis, the blood current being strong enough to penetrate the narrowed vessel, preventing thrombosis and necrobiosis. Thus we had only a transient interference with the function. The energetic antisyphilitic treatment commenced at once after this attack arrested the disease most effectually.

According to Heubner (*Dieluet. Erkrank. der Hirnarterien*, Leipzig, 1874, p. 186), the white matter of the cerebral base, even the chiasm, the cranial nerves, pons, and medulla are directly supplied by terminal branches of the large vessels in the same fashion as the central ganglia. Thrombosis in these branches from endarteritis obliterans must damage the area supplied by them. Marchand (*v. Graefe's Arch.*, xxviii., 2, p. 86) observed homonymous hemianopia from an infarction in one optic tract, and a case of Treitel and Baumgarten (*Virch. Arch.*, cxi., p. 251, 1888) is analogous. Treitel finds in this condition the best explanation of isolated or partial paralysis of cranial nerves. It is not necessary to find an atrophy in such instances, as the obstruction of the vessels need not be complete, so that only the function, not the nutrition, is abolished.

During life our first case exhibited two symptoms of affection of the visual path: homonymous hemianopia and papillitis. One of these, or optic neuritis, atrophy of the disc, temporal hemianopia, or other disturbances of the visual field was observed in more than 50 % of cases of cerebral lues. Papillitis and optic neuritis differ in this: a prominence of the optic disc of at least 2 D. (*i. e.*, 2/3 mm) is called papillitis, or less, optic neuritis. Both forms and simple atrophy are

about equally distributed (about 12 %). Papillitis is almost always double and, as in our case, most frequently caused by basal meningitis or gummata within the cranial cavity. In some instances it was the consequence of cerebral softening from syphilitic arteritis or of syphilitic periostitis of the orbit. Sometimes it makes the impression of a primary affection, when other cerebral symptoms are wanting, despite the actual existence of intracranial affections. It may disappear, without leaving traces, or may pass into atrophy, but the sight may recover. In our case it developed rather late in the series of cerebral symptoms and made the prognosis worse, as, in cerebral tumors, for instance, its occurrence precedes death by no long interval, if it sets in long after the symptoms of tumor.

Optic neuritis is also most frequent in basal gummous meningitis. Although it is to be considered as a descending neuritis, the intensity of its ophthalmoscopic appearance does not necessarily correspond to the intensity of anatomical changes in the optic nerve, especially when the cause of the visual disturbance lies farther back on the cranial base, as pointed out by Uhthoff. Thus there may be considerable failure of sight, ere slight changes of the disc are noticeable. According to Blessig (*Kl. M. f. A.*, 1875) it often is characterized (in comparison to papillitis) by rapid impairment of vision, less swelling, early grayish-white discoloration, and it may be one-sided. Sometimes stellate small white patches at the region of the macula are seen simultaneously. Primary syphilitic optic neuritis without other cerebral or ocular symptoms is rare. Atrophy of the optic disc, without preceding optic neuritis or papillitis, was observed in about 14 %, and it could always be proven to be secondary, quite differing from the primary gray atrophy of the optic nerve, *e. g.*, in tabes, by the peculiarity of the visual disturbances, especially the visual fields (Uhthoff, xxxix., 3, p. 157). It is mostly bilateral, complete or incomplete or partial, limited to the temporal half, as seen so often in amblyopia due to intoxication from tobacco and alcohol. Its aspect, however, has nothing characteristic of syphilis. Ophthalmoscopic changes of the retinal arteries of syphilitic origin, especially endar-



teritis, have been considered by Seggel, Ostwalt and others as premonitory symptoms of syphilis of the brain, others (Uthoff) hardly ever observed them in their cases of cerebral lues and doubt their intimate connection with the latter.

The functional disturbances in syphilitic affections of the visual path within the cranial cavity refer mostly to the field of vision, blindness being extremely rare. Homonymous hemianopia, as seen in two of our cases, is quite common and often of basal origin (in 4 out of Uthoff's 11 cases). If it is basal, as in our first case, it is due to an affection of one optic tract, and this is much more common in syphilis than in any other brain disease. It is marked by hemianopic reflex immobility of the pupils, by simultaneous affections of other cranial nerves, or by subsequent hemianopia of one or both remaining half-fields, when the chiasm becomes invaded, and by positive ophthalmoscopic changes of the optic discs, after a certain time, which generally consist in pallor. We had bilateral papillitis, seen four months after the first observation of hemianopia. It may have set in earlier, but I had not seen the patient for over a month. If he had lived longer, the papillitis might have turned into atrophy. According to literature temporal hemianopia occurs about twice as often (20 %) as homonymous (10 %) in syphilis of the brain, and is due to affection of the chiasm. If the visual disturbance is at first limited to one eye and temporal hemianopia sets in, the disease started in the optic nerve and spread to the chiasm. When commencing with homonymous hemianopia it originated in the optic tract and extended to the chiasm. In either case the non-decussating fibres of the other eye will be spared to the last, and before these are affected, the temporal hemianopia is one-sided. Later on there will be always ophthalmoscopic changes of the optic discs, optic neuritis or atrophy. Other forms of disturbances of the visual fields, as concentric limitations, central scotomata, enlargement of the blind spot or irregular defects with preservation of a peripheral crescent are occasionally observed and are mostly due to syphilitic disease of the optic nerve-trunks, generally associated with papillitis, ophthalmoscopically visible, but at first the latter may be wanting.

Total amaurosis in both eyes is very rare, and when observed it was mostly transient, yielding to antisymphilitic treatment.

This fact, as well as the form of the disturbance of the visual field, the oscillating course, and the complication with paralysis of other cranial nerves or parts of the brain are characteristic of the ocular symptoms in cerebral syphilis. Oppenheim explained this wavering of the visual complaints by the anatomical character of the syphilitic tissue: "This rapidly growing granulation-tissue exuberates and as rapidly decays in constant repetition, exposing the nerve invaded by it to ever-changing pressure."

The next frequent eye affection in syphilis of the brain is that of the ocular nerves. In Uhthoff's collection, out of 167 autopsies ( $150 + 17$ ) the optic fibres were diseased  $85 + 16 = 101$  times, the third nerves  $56 + 10 = 66$ , the sixth  $26 + 3 = 29$ , the fourth  $5 + 1 = 6$ , the fifth  $22 + 3 = 25$  times. Lesion of the third nerve, or both, is almost always basal, and is due to a gummous meningitis of the interpeduncular space or to gummata in this neighborhood. The gummous products not only act by pressure, producing atrophy, but infiltrate the nerve sheath and the nerve itself, leading to gummous perineuritis and neuritis. Or they develop more directly in the nerve, which degenerates, setting up syphilitic neuritis of the roots (Kahler), with swelling and proliferation of blood-vessels, or converting the nerve completely into gummous formations. Paralysis of one third nerve with crossed hemiplegia is due to gummata or foci of softening in the region of the peduncles or the pons, in consequence of thrombosis from syphilitic arteritis or from compression of the vessels at the base by the products of gummous meningitis.

This was observed in our *third case*:

A man, æt. thirty-two, had a primary sore 1891. March, 1893 paralytic seizure in bed (while travelling). A week later I saw him in consultation with his physician at his residence; *crossed hemiplegia, affecting limbs on right side, face on left, with total paralysis of left third nerve*. Ptosis, mydriasis, immobility of pupil, crossed diplopia, inability to read. V. in each =  $\frac{1}{3}$ . No ophthal-



moscopic changes. Tongue deviates to the right when put out; articulation slow and indistinct. Memory weak, has to think about words before he can utter them. He can walk, but very slowly. Antisyphilitic treatment without much result. I did not see him again until July, 1893, when he walked to my office: About the same condition. Ptosis and accommodation have improved. He reads No. 1, with either eye in 20 cm; pupil more than medium wide, contracts a little to light. April, 1894, ptosis entirely gone. Divergent squint; the left eye can be moved to the middle line, but not beyond. Pupil, 6.50 mm wide (R. 2.50 mm), reacts a little to light. He is not annoyed by double images, which he ignores. Psychically he is worse; he is not able to write a letter, forgetting what he wants to say. His left arm and leg tremble, and he can write his name tremulously only with great effort.

Diagnosis: *Syphilitic affection of left side of pons.*

When both third nerves are paralyzed from syphilitic new-formations in the interpeduncular space, it may happen that the most medial fibres are chiefly damaged. Although the posterior cerebral artery winds around the third nerve, it has not been found by Uhthoff in any of his cases causing oculomotor paralysis by compression or by thrombosis of its twigs supplying the nerve. From the fact that the affection of the third nerve in syphilis of the brain is mostly basal, we would infer that the paralysis observed during life would be total, according to the law that a partial ophthalmoplegia is either nuclear or fascicular, *i. e.*, where the nuclei and fibres of the different branches are separated. But anatomical researches have shown that apparently complete affections of the trunk of the third nerve at the base have caused only partial paralysis, viz., either ophthalmoplegia exterior or interior, isolated ptosis or paresis of only one or a few of the muscles supplied by the third nerve, or no symptoms at all as in our first case.

Our *fourth case* shows such a *partial paralysis of the third nerve of basal origin.*

A man, æt. thirty-seven, had a chancre thirteen years ago, and was treated for several years. Nine years ago he consulted some authority in Europe, who pronounced him cured, and consented to his marriage. He, his wife, and children did not show any

trace of syphilis during that time. Since December, 1893, he suffered from severe headache, but did not seek any medical aid. February, 1894, he suddenly felt sick, and was paralyzed (without losing consciousness), so that he could not walk any more. A week later I saw him, in consultation, on account of his eyes. Right hemiplegia, right ptosis, paresis of right internal rectus, and myosis. Right pupil 1 mm smaller than left. Crossed diplopia, inability to read with both eyes. Singultus since the attack lasting day and night for a week, disturbing his sleep and meals. Retention of urine in the first week. After six weeks the ptosis subsided, but returns a little towards evening, or when he is tired. No more double images. He goes out since that time, but his gait is staggering and uncertain. He complains of severe headache, mostly in occiput, but his mental condition is not much changed. Since April 1st his left arm has become weaker, which from the onset showed changes in the sense of temperature (all objects feeling warmer than the right), and his left leg has become ataxic.

The considerable variations of symptoms, the irregular palsies of limbs and cranial nerves (third) in this case, and the gradual deepening of the apoplectic seizure suggest a widely spread pathological process at the cerebral base. This may be either extensive basal syphilitic meningitis, injuring indirectly the vessels on the left, the third nerve on the right hemisphere, or syphilitic disease of the basilar artery with occlusion or incomplete obstruction of arteries that arise from it. The myosis may be due to an irritation of the pupillary fibres by the same process which paralyzed the other branches of the third nerve (analogous to Fontan's case, quoted by Mauthner, but explained as a nuclear affection).

In some cases of isolated ptosis the microscopical examination proved the nuclei to be healthy, but the nerve trunk diseased; a cortical lesion could never be found as cause (Uththoff xl., 1, pp. 63 and 67). The paralysis of the intrinsic or extrinsic muscles produced by a basal affection must be considered as an exception to the rule, which is, that it is nuclear or fascicular, but it may happen occasionally. Fascicular palsy of the third nerve was noticed in connection with crossed hemiplegia due to syphilitic disease in the pons or crus.



Paralysis of the sixth nerve, one or both, is in most instances basal from gummos meningitis, and mostly conjoined with palsies of other cranial nerves, or from endarteritis of the basilar artery with its sequelæ. If it is intracerebral, the lesion is in the pons. In none of the sixteen cases mentioned by Uhthoff could a nuclear affection be detected.

Our *fifth case* may illustrate this condition.

A man aged thirty-four had primary syphilitic infection, 1884. I saw him, in consultation, September, 1891. He had had eruption of skin, psoriasis plantarum, sores in mouth, pain in shin-bones. For about eight weeks he had headache with sleeplessness, and complained of his eyes. *Paralysis of right abducens.*  $V = \frac{1}{2}$  in each, no ophthalmoscopic changes. *Right homonymous partial hemianopia.* The hemianopic field is very irregular. Above and below it nearly reaches the median line (lacking  $10^{\circ}$ ). In some meridians it extends to the fixing point, but the periphery is partly preserved. This irregular defect is homonymous in both eyes. His headache commenced on the left side, and is now on the right. He is under great psychical depression, and cannot work. His memory of place (topographical ideation) has somewhat suffered, as he several times had difficulty to find his home, being confused about the streets. Antisyphilitic treatment improved his headache, but the paralysis of abducens did not disappear until summer, 1892. At that time his wife was confined with a child, who died with pemphigus after six months, according to the family physician, who also stated that the first child, five years old, is suffering from pemphigus. I did not see the patient any more until April, 1894. He is of healthy appearance and feels well, but has occasionally headache. Paralysis of abducens has entirely disappeared.  $V = \frac{1}{2}$ . Ophthalmoscopic condition normal, but the right partial homonymous hemianopia is just the same as at first.

*Diagnosis:* The absence of ophthalmoscopic changes of the discs after two years, the partial homonymous hemianopia, and the defective memory of place (Foerster, *v. Graefe's Arch.*, xxxvi., 1), would rather indicate a lesion of the visual centre in the left occipital lobe, than of the left optic tract, but the paralysis of the right abducens was due to an

affection in the right hemisphere. For these widely separated foci the best explanation would be, that a basal gum-mous meningitis injured on the right side mostly the nerves (abducens in this case), on the left the arteries (posterior cerebral), leading to thrombosis of some of its branches, which damaged portions of the occipital lobe supplied by them. Oppenheim (*Berl. klin. W.*, 1889, p. 1036) observed such conditions in hemiplegia with paralysis of cranial nerves of the same side. Our fourth cases with right hemiplegia and partial paralysis of right third nerve, is another instance of this category. Or it may be interpreted by an arteritis of the basilar artery, predominating in the left posterior cerebral and the right anterior inferior cerebellar branch. The analogy with Marchand's and Treitel's cases, mentioned above, would admit of a third explanation, viz.: The arterial disease may have settled in a terminal twig supplying the left optic tract, with consequent partial functional damage to the tract without following ophthalmoscopic changes. In that event, however, we might have expected a subsidence of the hemianopia as well, as the paralysis of the abducens ceased, the nerve fibres being more resistant than the brain substance of the centres. Thus, besides the other points mentioned, the recovery of the abducens, but the persistence of hemianopia, would be in favor of an affection of the occipital lobe and not of the optic tract.

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